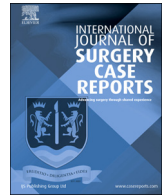




Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

## Spontaneous rupture of a giant renal angiomyolipoma—Wunderlich's syndrome: Report of a case

Panagiotis Nikolaos Chronopoulos<sup>a,\*</sup>, Georgios Nikolaos Kaisidis<sup>a</sup>,  
Christos Konstantinos Vaiopoulos<sup>a</sup>, Dragana Milosav Perits<sup>b</sup>,  
Michail Nikolaos Varvarousis<sup>c</sup>, Apostolos Vasilios Malioris<sup>d</sup>, Elissabeth Pazarli<sup>e</sup>,  
Ioannis Konstantinos Skandalos<sup>a</sup>

<sup>a</sup> Surgical Department, General Hospital, Kilkis, Greece<sup>b</sup> Anaesthesiology Department, General Hospital, Kilkis, Greece<sup>c</sup> Intensive Care Unit, Army Hospital 424, Thessaloniki, Greece<sup>d</sup> Urology Department, Army Hospital 424, Thessaloniki, Greece<sup>e</sup> Pathology Department, General Hospital "Papageorgiou", Thessaloniki, Greece

## ARTICLE INFO

## Article history:

Received 24 November 2015

Accepted 11 December 2015

Available online 17 December 2015

## Keywords:

Giant angiomyolipoma

Wunderlich's syndrome

Hypovolemic shock

Retroperitoneal hemorrhage

Nephrectomy

## ABSTRACT

**INTRODUCTION:** Herein we present a rare case of spontaneous rupture of a giant renal angiomyolipoma (AML), with symptoms of hypovolemic shock (Wunderlich's syndrome), which was managed by urgent total nephrectomy.

**PRESENTATION OF CASE:** A 53 year old female was transferred to the emergency room with progressive acute painful swelling of the left lateral abdominal area, duration of 5 h. An emergent ultrasonic examination, revealed a heterogeneous—solid mass with maximum diameter of 23 cm, with probable origin from the left kidney. Due to worsening of the clinical status (hypovolemic shock), loss of consciousness and acute drop of haematocrit level to 17.8%, the patient was urgently intubated in the emergency room and transferred to the operating theater. A giant haemorrhagic mass was found originating from the left kidney, which removed en-block with the left kidney. The patient was transferred to the intensive care unit. Her recovery was uneventful. The histopathologic examination revealed a giant renal angiomyolipoma (25 × 18 × 8 cm) with extensive bleeding.

**DISCUSSION:** Enlarged renal AMLs can rupture. This can be sudden and painful with manifestations of hypovolemic shock. The management of AMLs has been correlated with symptoms. Patients with life-threatening retroperitoneal haemorrhage, require urgent exploration as retroperitoneal bleeding can lead to severe complications, increasing morbidity.

**CONCLUSION:** In case of giant angiomyolipoma with intratumoral haemorrhage, and symptoms of Wunderlich's syndrome, partial or total nephrectomy is a good treatment option in order to save the patient's life.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Renal angiomyolipoma (AML) is an infrequent tumor that, in most cases, follows a benign course and has clearly defined radiological [1] and histological characteristics [2]. Enlarging AMLs can develop micro and macro-aneurysms that can rupture. This can be sudden and painful, and occasionally life threatening. In 10% of the cases, they manifest as hypovolemic shock, resulting from massive retroperitoneal bleeding, known as Wunderlich's syndrome

[3]. The frequency of symptoms and the risk of bleeding due to rupture increases with the size of AML.

We present a rare case of hypovolemic shock due to rupture of a giant renal AML (Wunderlich's syndrome), which was managed by urgent total nephrectomy.

## 2. Presentation of case

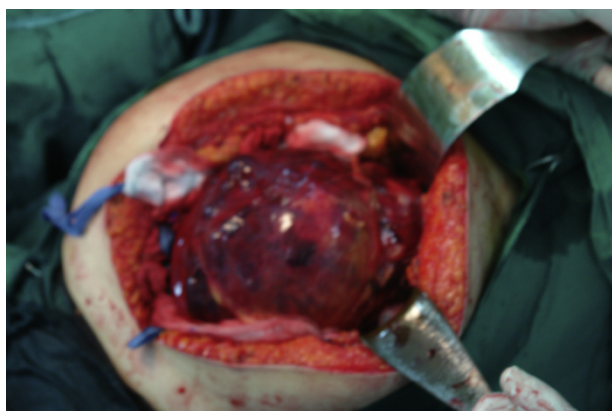
A 53 year old female was transferred to the emergency room of our provincial hospital with symptoms of acute left abdominal pain, duration of 5 h, accompanied with progressive painful swelling of the left lateral abdominal area (Fig. 1). Her past medical history was free. Upon clinical examination, a hard painful swelling was noted along her left abdomen as well as symptoms

\* Corresponding author.

E-mail address: [pachrono@gmail.com](mailto:pachrono@gmail.com) (P.N. Chronopoulos).

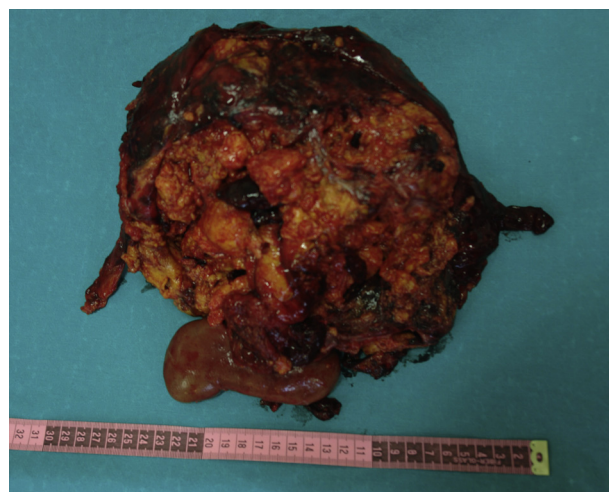


**Fig. 1.** Swelling of the left lateral abdominal area.



**Fig. 2.** Disruptions of the retroperitoneum and a pulseless, sizeable, abnormal bleeding mass occupied all of the left retroperitoneal area.

of paleness, sweating and tachycardia. Arterial pressure was 75/50 mmHg, pulse 120 per min. An emergent ultrasonic examination, revealed a heterogeneous—solid mass separated by diaphragms, with maximum diameter of 23 cm, in the left abdominal area, with probable origin from the left kidney. Computer tomography scanning was not performed due to non-functioning of the hospital's CT scanner. Original blood analysis showed Hct 23% and Hb 7.6 mm/dl. Due to worsening of the clinical status and loss of consciousness, the patient was urgently intubated in the emergency room and transferred to the operating theater. Her Hct fell to 17.8%. An abdominal midline incision was made extending from the xiphisternum to just above the pubic symphysis. Free blood was found in the peritoneal cavity, disruptions of the retroperitoneum and a pulseless, sizeable, abnormal bleeding mass occupied all of the left retroperitoneal area (Fig. 2). The left posterior parietal peritoneum was transected and the abdominal aorta was checked blindly up to the level of the renal arteries. The giant haemorrhagic mass was found originating from the left kidney, was mobilized and removed en-block with the left kidney (Fig. 3). The retroperitoneal area was washed, a haemostasis was done and the parietal peritoneum was sutured. Drainage was placed in the left coloparietal region, also the lesser pelvis and the abdominal wall was closed. 4 blood units were transfused. The intubated patient was transferred to the intensive care unit of another civil hospital.



**Fig. 3.** The surgical specimen: Giant haemorrhagic mass (25 × 18 × 8 cm) originated from the left kidney, en-block with the left kidney.

Her recovery was uneventful. The patient was extubated on the second postoperative day and she was discharged from the hospital on the 8th post-operative day.

The histopathologic examination revealed a giant angiomyolipoma originating from the left kidney (25 × 18 × 8 cm) with extensive bleeding (Fig. 4a and b). Subcapsular haemorrhagic infiltrations and scarring of glomeruli in the region of the tumor outgrowth were found in the renal parenchyma (6.5 × 3.5 × 2.5 cm). No evidence of malignancy.

Routine biochemistry and blood analysis at 4 months post-operative follow-up were normal. Abdominal computer tomography did not show tumor recurrence or other pathological findings in the right kidney as well as in other organs.

### 3. Discussion

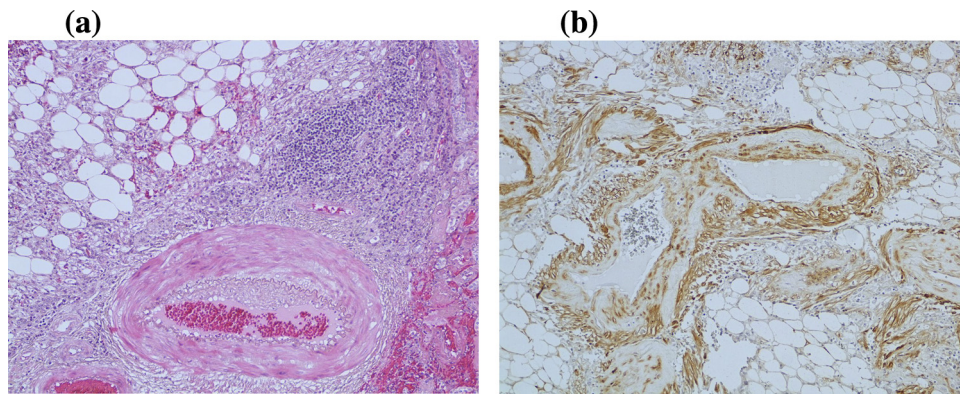
The AMLs are uncommon tumors. They were first described in 1951 by Morgan et al. [4]. Most of these tumors appear in the kidney, however these can be found in other locations, such as the spleen, liver, uterus and fallopian tubes [5,6].

Renal AML is a benign tumor known to occur sporadically and in association with genetic syndromes like tuberous sclerosis (TS) and lymphangioleiomyomatosis. Patients with lymphangioleiomyomatosis may have multiple renal and hepatic AMLs, multiple pulmonary cysts, enlarged abdominal lymph nodes and lymphangiomyomas [6,7]. Renal AMLs arise from the mesenchymal elements of the kidney and they are composed of varying proportions of mature adipose tissue, smooth muscle and abnormally thick-walled blood vessels [8]. For this reason they are also known as renal hamartomas.

AMLs are found in approximately 45–80% of patients with TS and tend to occur in adolescents and young adults. Tuberous sclerosis is a familiarly inherited disorder comprising adenoma sebaceum, mental retardation and epilepsy [9]. Bilateral renal AML in TS is a rare entity and the female/male prevalence is nearly equal [10].

The overall prevalence of AMLs, in population, is 0.44% with a pronounced female predominance (2:1) [11], suggesting a hormonal component to tumor growth [6]. In most patients it presents later in life, during the fifth or sixth decade, as in our present case.

Most small AMLs are asymptomatic and found incidentally upon radiological studies [6]. The classical clinical presentation of AML is a palpable tender mass, flank pain and gross haematuria, known as Lenk's triad [12]. Less frequent associated symptoms include nausea or vomiting, fever, anaemia and blood pressure alteration.



**Fig. 4.** (a) (HE  $\times 10$ ) Renal angiomyolipoma with haemorrhagic infiltrations. (b) (SMA  $\times 10$ ) Fascicles of smooth muscle cells.

The two major morbidities associated with renal AML are retroperitoneal haemorrhage and impingement of AML on the kidneys and other vital organs [6]. Enlarged AMLs can develop micro and macro-aneurysms that can rupture. This can be sudden and painful and occasionally life threatening. In 10% of the cases, they manifest as hypovolemic shock resulting from massive retroperitoneal bleeding, known as Wunderlich's syndrome [3,12]. The frequency of symptoms and the risk of bleeding (rupture) increases with the size of an AML [13].

Rupture can occur spontaneously in patients on anticoagulants, or with trauma to the kidney. Rupture has also been reported during pregnancy or in the post-partum period [6].

AML is the only benign renal tumor that is confidently diagnosed on cross-sectional imaging. There are two imaging characteristics that are highly suggestive of an angiomyolipoma. On ultrasound, they are echogenic. On CT scanning, they have the density of fat, which is less than that of water [14]. The presence of fat (confirmed on non-enhanced thin-cut computed tomography) by a value of  $-20$  Hounsfield units or less seen within a renal lesion on imaging is considered the diagnostic hallmark. MRI can be used to identify the fatty tissue. However because the presence of bleeding in any renal tumor can mimic the typical pattern of angiomyolipoma, MRI should not be considered the diagnostic method of choice. AML can vary in size from a few millimeters to larger than 20 cm. It is unusual to see an AML over 10 cm, therefore many studies have demonstrated that any AML measuring over that number is considered as "giant". CT findings usually help to differentiate AMLs from other tumors, such as perinephric liposarcomas [1].

Arterial angiography can reveal neovascularity, similar to that of renal cancer and therefore it is not helpful in differential diagnosis.

The management of AMLs has been correlated with symptoms. Most patients with small tumors ( $<4$  cm) that tend to be asymptomatic are managed conservatively, under periodic ultrasonography follow up [13]. Radiofrequency ablation (RFA) of renal tumors has been proven as a useful treatment for small, incidentally found, renal cortical lesions in elderly patients or patients with a solitary kidney, at high risk of renal dysfunction, following partial nephrectomy [15].

Tumors larger than 8 cm generally tend to be symptomatic and they are at a greater risk of spontaneous or traumatic rupture resulting in haemorrhagic complications. These patients are therefore treated by angiography and selective arterial embolisation as a first line [14]. Prophylactic embolisation of asymptomatic lesions 4 cm or larger is recommended in selected high risk patients, including younger women who intend future pregnancy or in patients in whom regular follow-up is difficult. Embolisation reduces the risk of haemorrhage by blocking the blood supply to the AML. The tumor size usually decreases by two-thirds up to 40%. Recurrence

of the tumor has been reported and repeated embolisations are occasionally needed.

Partial or radical nephrectomy is indicated, if there is persistent haemorrhage, suspicion of malignancy or failed embolisation. In cases of bilateral lesions, as seen in TS, nephron-sparing surgery (by either selective embolization or open or laparoscopic/robotic partial nephrectomy) must be performed [16]. Patients with life-threatening haemorrhage, require urgent exploration and in the majority of the cases require total nephrectomy [17]. We considered nephrectomy since our patient was having acute drop of haematocrit levels, increased size of retroperitoneal haematoma and hypovolemic shock (Wunderlich's syndrome).

Histopathologically, AML consists of mature adipocytes, thick-walled blood vessels, and epithelioid stromal cells in various proportions [8]. Usually it displays as a pattern of typical fat and perivascular epithelioid cells arranged around a blood vessel. Positive immunoreactivity for HMB-45, a monoclonal antibody raised against a melanoma-associated antigen, is characteristic for AML and can be used to differentiate this tumor from sarcoma and other tumors [6].

#### 4. Conclusion

Patients with a life-threatening angiomyolipoma haemorrhage, require urgent intervention, as retroperitoneal bleeding can lead to severe complications, increasing morbidity. In case of giant angiomyolipoma with intratumoral haemorrhage, and symptoms of Wunderlich's syndrome, partial or total nephrectomy is a good treatment option in order to save the patient's life.

#### Conflict of interest

The author declares no conflict of interest.

#### Funding

Nothing to declare.

#### Ethical approval

Nothing to declare.

#### Consent

Nothing to declare.



**Author contribution**

Nothing to declare.

**Guarantor**

Nothing to declare.

**References**

- [1] N. Schieda, A.Z. Kielar, O. Al Dandan, M.D. McInnes, T.A. Flood, Ten uncommon and unusual variants of renal angiomyolipoma (AML): radiologic–pathologic correlation, *Clin. Radiol.* 70 (2015) 206–220.
- [2] T.A. Skolarus, M.F. Serrano, D.A. Berger, T.L. Bullock, Y. Yan, P.A. Humphrey, A.S. Kibel, The distribution of histological subtypes of renal tumors by decade of life using the 2004 WHO classification, *J. Urol.* 179 (2008) 439–443, discussion 443–444.
- [3] Y.C. Chen, Y.C. Lin, Wunderlich syndrome, *QJM* 106 (2013) 187–88.
- [4] G.S. Morgan, J.V. Straumfjord, E.J. Hall, Angiomyolipoma of the kidney, *J. Urol.* 65 (1951) 525–527.
- [5] D.E. Maziak, S. Kesten, D.C. Rappaport, J. Maurer, Extrathoracic angiomyolipomas in lymphangioleiomyomatosis, *Eur. Respir. J.* 9 (1996) 402–405.
- [6] J.N. Eble, Angiomyolipoma of kidney, *Semin. Diagn. Pathol.* 15 (1998) 21–40.
- [7] Z.F. Peng, L. Yang, T.T. Wang, P. Han, Z.H. Liu, Q. Wei, Efficacy and safety of sirolimus for renal angiomyolipoma in patients with tuberous sclerosis complex or sporadic lymphangioleiomyomatosis: a systematic review, *J. Urol.* 192 (2014) 1424–1430.
- [8] B. Kulkarni, S.B. Desai, B. Dave, H.B. Tongaonkar, J.N. Kulkarni, R.F. Chinoy, Renal angiomyolipomas—a study of 18 cases, *Indian J. Pathol. Microbiol.* 48 (2005) 459–463.
- [9] P.F. Bolton, M. Clifford, C. Tye, C. Maclean, A. Humphrey, K. le Maréchal, J.N. Higgins, B.G. Neville, F. Rijdsdijk, Tuberous Sclerosis 2000 Study Group, J.R. Yates, Intellectual abilities in tuberous sclerosis complex: risk factors and correlates from the Tuberous Sclerosis 2000 Study, *Psychol. Med.* (April) (2015) 1–11.
- [10] C. Bansal, V. Lakshmaiah, A. Raavsha, P. Waadhawan, M.K. Priyanka, Tuberous sclerosis with bilateral renal angiomyolipoma, *J. Assoc. Physicians India* 61 (June (6)) (2013) 420–423.
- [11] A. Fittschen, I. Wendlik, S. Oeztuerk, W. Kratzer, A.S. Akinli, M.M. Haenle, T. Graeter, Prevalence of sporadic renal angiomyolipoma: a retrospective analysis of 61,389 in- and out-patients, *Abdom. Imaging* 39 (2014) 1009–1013.
- [12] A. Guttilla, A. Crestani, F. Cattaneo, F. Zattoni, C. Valotto, M. Iafrate, F. Dal Moro, F. Zattoni, Wunderlich's syndrome: three cases of acute spontaneous renal bleeding conservatively treated, *Arch. Ital. Urol. Androl.* 85 (2013) 210–213.
- [13] S. De Luca, C. Terrone, S.R. Rossetti, Management of renal angiomyolipoma: a report of 53 cases, *BJU Int.* 83 (3) (1999) 215–218.
- [14] A. Bora, A. Soni, N. Sainani, D. Patkar, Emergency embolization of a bleeding renal angiomyolipoma using polyvinyl alcohol particles, *Diagn. Interv. Radiol.* 13 (2007) 213–216.
- [15] S.M. Gregory, C.J. Anderson, U. Patel, Radiofrequency ablation of large renal angiomyolipoma: median-term follow-up, *Cardiovasc. Intervent. Radiol.* 36 (2013) 682–689.
- [16] S. Faddegon, A. So, Treatment of angiomyolipoma at a tertiary care centre: the decision between surgery and angioembolization, *Can. Urol. Assoc. J.* 5 (2011) E138–E141.
- [17] A. Granata, A. Basile, M. Figuera, R. Mignani, C.E. Fiore, Spontaneous retroperitoneal due to massive rupture of renal angiomyolipoma treated with nephrectomy: an unusual onset of tuberous sclerosis complex, *Clin. Nephrol.* 71 (2009) 441–444.

**Open Access**

This article is published Open Access at [sciedirect.com](http://sciedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.